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Independence and Adaptive Behavior in Adults With Williams Syndrome

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This study describes the adjustment of 70 adults with Williams syndrome, in terms of self-help skills, independence, and occupational levels. Although the overall mean IQ of the group (62.00) was within the mild mental handicap range, relatively few individuals were able to attain a high level of independence or cope with the demands of employment. Adaptive behavior scores were significantly below chronological age. Outcome measures were compared with available data on other groups of adults of similar age and level of intellectual impairment. Implications for the community care of adults with Williams syndrome are discussed. Am. J. Med. Genet. 70:188–195, 1997. © 1997 Wiley-Liss, Inc.

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INTRODUCTION

Williams syndrome (WS) is a genetically determined, intellectually disabling condition, which occurs in approximately 1 in 25,000 live births [Jones and Smith, 1975]. Most cases occur sporadically, although there are a few accounts of parent-to-child transmission [Morris et al., 1993]. A microdeletion on chromosome 7 has been identified in affected individuals, occurring around the site of the elastin gene [Ewart et al., 1993].

The characteristic phenotype of WS in childhood is now well-documented. The condition typically presents in infancy, with feeding difficulties, profuse vomiting, irritability, constipation, and poor weight gain [Martin et al., 1984]. There is a distinctive facial appearance, with full prominent cheeks, full lips and a wide mouth, a long philtrum, an "upturned" nose with flat nasal bridge, prominent orbital ridges, medial eyebrow flare, and stellate iris pattern [Joseph and Parrot, 1958]. A subgroup of affected infants presents with idiopathic infantile hypercalcemia, which can be effectively treated by dietary intervention.

A number of cardiovascular abnormalities has been described in WS, notably supravalvular aortic stenosis and peripheral pulmonary artery stenosis [Hallidie-Smith and Karas, 1988]. Renal problems are also common, in particular renal artery stenosis and renal hypertension, which are reported to worsen with age [Ingelfinger and Newburger, 1991; Pober et al., 1993]. Other findings include strabismus, dental abnormalities, and growth retardation.

WS is accompanied by cognitive delays. In a study of affected children of school age, Udwin et al. [1987] found that 95% had moderate to severe mental handicap, while 5% had borderline cognitive abilities. Language may be initially delayed, but typically develops to a level that is markedly superior to visuospatial abilities and motor skills [Udwin and Yule, 1990, 1991]. In most cases, expressive language skills are relatively good: speech is grammatically correct, complex, and fluent at a superficial level, but also verbose and pseudomature. Comprehension is often more limited than expressive language suggests, and excessive and frequently inappropriate use of clichés and stereotyped phrases is typical. Children with WS tend to be very chatty, with a precocious vocabulary, and good use of social language and mimicry skills. Auditory memory and verbal processing skills are particularly well-developed. In contrast, fine and gross motor coordination, numeracy, and the ability to integrate visuospatial information are frequently poorly developed [Bellugi et al., 1994].

Children with WS also appear to have a distinctive emotional and behavioral profile, including overactivity and poor concentration, attention-seeking behavior, excessive anxiety, hyperacusis, preoccupations and obsessions, eating and sleeping difficulties, and poor peer relationships. Most tend to be socially disinhibited and overfriendly in interactions with others [Udwin et al., 1987]. Udwin et al. [1987] found higher rates of behavioral disturbance in these children in comparison with other groups of children of similar cognitive ability.

Despite the large body of data on affected children,

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there is little systematic information on the long-term progression of the condition. Morris et al. [1988, 1990], Kaplan et al. [1989], and Ingelfinger and Newburger [1991] noted progressive medical problems in small numbers of WS adults, with a high frequency of hypertension, cardiovascular and gastrointestinal problems, urinary tract abnormalities, and progressive joint limitations. In contrast, Lopez-Rangel et al. [1992] and Plissart et al. [1994] found few serious health problems in their subjects, who were age 18–66 years. There is a suggestion that individuals with WS age prematurely, and greying hair and a "coarse" facial appearance are common even in the early to midtwenties.

Other studies have pointed to the lack of independence and the high levels of support required by adults with WS. In a survey of 119 adults from age 16–38 years, Udwin [1990] found that the pattern of psychological and personality characteristics identified in children with WS persists virtually unchanged into adulthood. Despite their relatively good verbal and social skills, most of the adults were unable to live independently and required ongoing support and supervision in everyday activities. Most attended day care centers or had no daytime occupation. In an earlier study, Martin et al. [1984] collected preliminary data on 25 affected adults, and noted that only 4 were in paid employment and 3 attended job-training schemes. The remaining 18 attended training centers for adults with cognitive disability.

There have been no other detailed investigations of individuals with WS as they grow into adulthood, and little is known about their levels of independence in comparison with other groups of individuals with similar levels of cognitive disability. We carried out detailed assessments and interviews with adults with WS, their parents, caregivers, and employers, and in this paper we describe level of independence, living arrangements, and occupational activities of the group, and compare the results with findings obtained in similar studies of adults with other intellectually disabling genetic conditions.

SUBJECTS AND METHODS

Subjects

The subjects were drawn from past and present membership of the Williams Syndrome Foundation, a parent support group which holds the only register of affected individuals in the United Kingdom.

Preliminary postal questionnaires were sent to the parents (or other caregivers) of 207 individuals known to the Foundation who were age 18 years or older on January 1, 1994. The questionnaires sought the families' agreement to participate in the study, as well as brief information concerning diagnosis, current living arrangements, daytime occupations, educational attainment, and self-care skills. A total of 185 families (92%) responded, and 182 (90%) agreed to participate in the interviewing phase of the study. Fifty-four of these were excluded from the second phase of the study because there was insufficient information to confirm diagnosis. The remaining 128 adults had all previously been diagnosed as having WS (or idiopathic infantile hypercalcemia) by pediatricians, clinical geneticists, and/or cardiologists on the basis of their medical history, facial anomalies, and a physical examination. At the time the study was undertaken (1994–1995), elastin deletion studies were not widely available, and none was undertaken for the purpose of confirming the diagnosis. All subjects had learning difficulties, and many were confirmed to have supravalvular aortic stenosis or peripheral pulmonary artery stenosis. Those cases where an unspecified cardiac anomaly (e.g., "heart murmur") was recorded were included only if infantile hypercalcemia was present in childhood. These inclusion criteria accord with those set out by Martin et al. [1984] for a diagnosis of WS. The average age at initial diagnosis was 3 5/12 years (SD 60.9 months; range, 5 months–24 years).

From the information obtained in the initial postal questionnaires, each subject was rated by two of the authors (M.D. and O.U.) as having borderline, moderate, or severe mental retardation and as having high, moderate, or low levels of independence. Using these ratings, a representative sample of 62 subjects was selected to take part in the interview stage of the study. In addition, because reports of high-functioning adults with WS are rare, a further 8 subjects who scored high on subjective measures of independence, or were rated as being of borderline cognitive ability, were also included in the interview phase. Thus, it should be noted that the final group of 70 subjects was on average more able than the population of adults with WS in general (see Table I).

Methods

Each of the 70 subjects was assessed on the following battery of cognitive and educational measures: the Wechsler Adult Intelligent Scale-Revised (WAIS-R) [Wechsler, 1986]; the British Picture Vocabulary Scale-Short Form [Dunn et al., 1982]; Expressive One-Word Picture Vocabulary Test [Gardner, 1979]; Wechsler Objective Reading Dimensions [Wechsler, 1993]; and Raven's Progressive Coloured Matrices [Raven, 1956]. Details of the cognitive functioning of adults with Willi-

TABLE I. Sample Characteristics

		N = 70[a]		N = 128[b]	
		n	%	n	%
Gender	Male	33	45	58	45
	Female	37	55	70	55
Cognitive ability	Borderline	15	21	15	12
	Moderate	27	39	53	41
	Severe	28	40	60	47
Independence	High	9	13	9	7
	Moderate	30	43	52	41
	Low	31	44	67	52
Age distribution (years)	18–24	32	46	66	52
	25–29	21	30	39	31
	30–34	8	11	11	9
	35–39	9	13	11	9

[a]Subjects selected from those families who responded to the postal questionnaire.
[b]Respondants to the postal questionnaire where the diagnosis of WS was confirmed.

ams syndrome are presented in more detail by Howlin et al. [1997].

In addition, the parents and caregivers of each adult were interviewed by one of the authors (M.D.). Interviews with the parents and caregivers each took approximately 3 hr to complete, and sought information on the adults' health, living arrangements, self-care and daily living skills, educational attainment, daytime occupations since leaving school, extent of support received from professional agencies, social relationships, and behavioral and emotional difficulties. The caregivers' responses were used to complete the Vineland Adaptive Behavior Scale-Interview Edition [Sparrow et al., 1984] in each case. Sixty-six subjects were interviewed; the remaining 4 adults did not wish to undertake an interview themselves, but gave their consent for their parents and supervisors to provide information. The interview with each adult took approximately 1 hr to complete, and sought information about their satisfaction with current living arrangements; satisfaction with further education and training courses undertaken; and satisfaction and problems encountered in daytime occupations. For each subject with a daytime occupation (N = 59), the relevant employer, college tutor, training center supervisor, or keyworker also was interviewed. These interviews sought information on the subjects' abilities and the difficulties they encountered in their daytime occupations and activities.

RESULTS

Cognitive Ability and Vineland Adaptive Behavior Scores

The sample had a mean chronological age of 26 years, 8 months (SD 68.5 months; range, 19 years–39 years, 9 months). The mean full-scale IQ of the group, as assessed on the WAIS-R, was 62.00 (see Table II). Most of the subjects (84%) obtained IQs in the range of 50–70, 8 (11%) obtained an IQ in the range of 71–80, and 3 (4%) obtained IQs below 50.

Table III presents the scores on the Vineland Adaptive Behavior Scales. These scores suggest that the group was stronger in certain aspects of adaptive behavior than in others. Written communication, play and leisure, coping skills, and community skills appear to be areas of relative strength, while the group attained lower scores on receptive communication skills and interpersonal relationships. However, the highest age equivalent score possible on the receptive communication subdomain of the Vineland Adaptive Behavior Scale is 94 months; it should be noted therefore that low attainment in this area may be due to an artifact of the scale used.

TABLE II. Full-Scale IQ, Verbal IQ, and Performance IQ Means, Standard Deviations (SD), and Ranges

	Mean	SD	Range
Full-scale IQ	62.00	6.76	46–84
Verbal IQ	65.76	6.86	52–91
Performance IQ	61.57	5.80	51–79
Chronological age (months)	320.19	68.54	228–477

Correlations between full-scale IQ and standard scores on each of the three Vineland domains were all positive and significant (see Table IV). The correlations between IQ and both the Daily Living Skills and Communication domains were higher than between IQ and the Socialization domain.

Self-Care, Domestic Skills, and Leisure Activities

Table V presents caregivers' assessments of the amount of supervision required by the subjects to perform a variety of self-care tasks and daily chores. About a quarter of the subjects required assistance or supervision when using the toilet, and about half required at least some assistance with washing and dressing. Between 80–94% were wholly dependent on others for preparation of food and domestic chores such as cleaning, shopping, and laundry. Few subjects could tell the time or use the telephone competently, and only one person could use money appropriately, and controlled her own finances, bank accounts, and expenditures. In all the remaining cases, the caregivers took responsibility for budgeting and buying essential items, although 3 subjects could shop independently.

No subject could drive independently, although 5 (7%) had received driving lessons. Parents reported that these individuals were extremely anxious while driving, even under supervision; they had poor coordination, difficulties judging speed and distance, and became anxious at junctions and when meeting oncoming traffic. Although 28 of the group (40%) were able to use public transport on familiar journeys unaccompanied, only 2 were able to use public transport to make unfamiliar journeys. Caregivers reported that the majority of subjects could read and write, at least at a simple level. However, only 13 (19%) read adult newspapers or novels, while only 3 (4%) were considered capable of writing a formal letter.

Relatively few subjects (6%) had a range of leisure interests in which they participated regularly. Nine (13%) had some spontaneous leisure interests, but these were of limited scope or frequency. Forty-five (64%) participated in activities only if they were arranged by others, and 2 subjects had no leisure activities at all. However, 34 (49%) attended organized clubs on a weekly basis, and 6 (9%) attended clubs less frequently. The majority (68%) of those who regularly attended clubs encountered no problems, while the remaining 32% frequently had problems getting on with others. Many parents reported that their son or daughter enjoyed swimming, but only 12 of the group (17%) played sports, and it was noted that as individuals got older they became less willing to participate in activities of this kind. Eight subjects (11%) could play keyboards or percussion instruments, and a further 43 (61%) were reported to have musical ability that was better than their general level of functioning.

The majority of the group (87%) were reported to have circumscribed interests and preoccupations of unusual intensity. A number of subjects shared interests

TABLE III. Mean Age Equivalent Scores and Standard Deviations for Domains and Subdomains of the Vineland Adaptive Behavior Scale

	Mean age (months)	SD	Range (months)	Maximum age equivalent score possible (months)
Communication	82.27	33.63	40–228	228
Receptive	58.79	22.15	30–94	94
Expressive	75.43	33.77	23–186	186
Written	90.97	34.97	49–228	228
Daily living skills	79.63	37.17	25–228	228
Personal	75.77	41.58	18–210	210
Domestic	73.37	32.15	16–228	228
Community	86.11	41.69	24–228	228
Socialization	81.10	42.06	17–228	228
Interpersonal relationships	70.74	48.05	8–228	228
Play and leisure	90.99	48.86	11–228	228
Coping skills	86.77	43.98	10–228	228

in similar topics such as power tools, cars, animals, the royal family, and television or film personalities. However, only six families reported that such interests interfered with daily life.

Living Arrangements

Living arrangements are shown in Table VI. The majority of subjects still lived at home with their families, and only 3 were living independently. Of the 48 families whose son or daughter lived at home, 29 (60%) reported being "very satisfied" with these living arrangements. Of those who expressed a degree of dissatisfaction, eight families thought that the adult would be better off living in sheltered accommodation, and nine thought that a group home would be preferable. Sixty percent of the families who were the primary caregivers expressed significant concerns about future difficulties, mainly concerning their continuing ability to look after their son or daughter over time, and concerning the individual's lack of social and personal independence. Three families (6%) reported that they were already having problems coping with the needs of their son or daughter.

Twenty-two subjects (31%) had left the parental home. Of these, one was married and lived independently with her husband, and 2 others lived in independent accommodation. In these 3 cases, the parents organized and financed the living arrangements, and continued to provide practical support. In each case, the parents mentioned that the subjects had problems coping with money and domestic chores, and two of the families would have preferred a sheltered, supervised residential setting for their son or daughter. However, in all 3 cases the subjects themselves reported no problems coping with living independently, and none wished to live in an alternative setting.

The remaining 19 subjects who had left the parental home lived in residential or educational units. Fourteen of these families expressed satisfaction with this accommodation. However, five families expressed concerns about inadequate supervision, residences that were very busy or noisy, or settings which were subject to frequent changes of staff or where the individual was frequently moved from one residence to another.

TABLE IV. Mean Standard Scores and Standard Deviations for Adaptive Behavior Domains and Their Correlation (r) With Full-Scale IQ

	Mean	SD	r	P
Communication	34.17	19.26	0.70	< .001
Daily living skills	36.53	19.42	0.57	< .001
Socialization	44.61	17.65	0.38	< .001

Daytime Occupations

Details of the subjects' daytime occupations are given in Table VII.

Employment. Only one person had an independent job, as part-time kitchen assistant in a large department store. Her supervisor reported that she was having substantial problems coping with the strict hygiene standards required, as well as with the general demands of the job, such as timekeeping and coping with the heat of the kitchens. She also found it difficult to get on with some of her colleagues. Four subjects worked in sheltered employment: 2 as supermarket assistants, 1 as a kitchen assistant, and 1 as an assembly line packer. Five people had part-time voluntary jobs: 3 helped in old peoples' homes; 1 had a voluntary job as an office assistant; and the other was a kitchen assistant. A further 11 individuals were undertaking part-time work placements organized by their adult train-

TABLE V. Self-Care and Domestic Skills

	Independent	Needs assistance	Dependent
	n (%)	n (%)	n (%)
Toileting	51 (73%)	12 (17%)	7 (10%)
Washing	39 (56%)	20 (29%)	11 (16%)
Dressing	30 (43%)	25 (36%)	15 (21%)
Clothes care	1 (1%)	5 (7%)	64 (92%)
Cooking	3 (4%)	11 (16%)	56 (80%)
Cleaning	2 (3%)	6 (9%)	62 (89%)
Shopping	1 (1%)	3 (4%)	66 (94%)
Timekeeping	11 (16%)	18 (26%)	41 (59%)
Budgeting finances	1 (1%)	3 (4%)	66 (94%)
Telephone skills	9 (13%)	44 (63%)	17 (24%)
Driving	0 (0%)	5 (7%)	65 (93%)
Public transport	2 (3%)	28 (40%)	40 (57%)
Reading	13 (19%)	26 (37%)	31 (44%)
Writing	3 (4%)	40 (57%)	27 (39%)

TABLE VI. Living Arrangements

	n	(%)
At home with relatives	48	(69%)
Hostel or group home	8	(11%)
Sheltered accommodation (some staff support)	7	(10%)
Independent accommodation	3	(4%)
Residential further education college	2	(3%)
Residential community	2	(3%)

ing center or further education college. Such placements were for 1 or 2 days a week, and included jobs such as kitchen assistant, shop or supermarket assistant, packer, and nursery helper.

The principal daytime supervisors of the 21 subjects with jobs and work placements were interviewed about the abilities and difficulties of the WS adults in the workplace. In half of these cases, supervisors reported that subjects were having problems which "threatened their employment," and a further one third were said to be experiencing difficulties which required "additional supervision." The main problems encountered in the various work settings are summarized in Table VIII.

Further education. One third of the group (23) was following a course at a further education college (see Table VII). These courses typically cater to individuals with special needs who have left school but require additional education and independence training, and usually last for 2 years. Further education courses were provided by both residential and community day-colleges. A further 28 subjects had taken a further education course prior to taking part in this study. Of this total group of 51 individuals, 36 families (69%) said they were largely satisfied with the further education received, while 16 families and caregivers (31%) were largely dissatisfied. The main reasons cited for dissatisfaction were inappropriate courses or training (e.g., teaching bricklaying to someone with poor motor skills), lack of supervision, and lack of feedback to parents. Thirty-three parents (65%) reported that their son or daughter had experienced difficulties while at college, such as difficulties coping with coursework (4 subjects); difficulties with timekeeping and attendance (2 subjects); interpersonal difficulties (12 subjects); and a combination of these (13 subjects). Parents of 2 subjects reported problems but were unable to explain what these were. The problems associated with attending further education were very similar to those reported by employers: anxiety, distractibility, inappropriate social behaviors, and motor difficulties.

Adult centers. Twenty-six (37%) of the sample attended day centers of some kind. These centers are funded by local social services and provide day care for individuals with a wide range of physical and intellectual disabilities. However, they varied greatly in the nature and quality of provision offered; some provided vocational and independence training, while others tended to provide leisure activities for adults with more severe impairments. Twenty-five subjects attending such centers were willing to be interviewed. Seventeen were "very satisfied" with the activities provided by their center; the remaining 8 adults reported interpersonal problems and "boredom" as reasons for dissatisfaction. Four individuals had part-time work placements, and 11 others were taught vocational skills. Activities undertaken in the centers included educational activities (11 subjects), lessons in daily living skills such as using public transport and coping with money (14 subjects), and arts and crafts activities (13 subjects). All but 7 individuals said they took part in leisure and social activities during the week. Twenty-three parents and caregivers were satisfied that the placement was appropriate for the needs of the individual, although lack of appropriate activities, high staff turnover, and poor communication with families were mentioned by the remainder. Two adults lived in residential communities, and had no daytime occupation other than community activities. Nine subjects living at home had no formal daytime occupation at the time of the study.

TABLE VII. Daytime Occupations

	n	(%)
Independent employment	1	(1%)
Sheltered employment	4	(6%)
Voluntary work	5	(7%)
No daytime occupation	11	(16%)
Further education	23	(33%)
Adult training center/day center	26	(37%)

TABLE VIII. Problems Encountered by Adults With WS With Jobs and Work Placements (n = 21)

	n	(%)
Overfriendliness	21	(100%)
Anxiety	19	(90%)
Distractibility	19	(90%)
Excessive or inappropriate chatter	18	(86%)
Dislike of changes in routine	15	(71%)
Intolerance of others	12	(57%)
Anger management	8	(38%)
Fine motor difficulties	8	(38%)
Gross motor difficulties	7	(33%)
Personal appearance/hygiene	7	(33%)

Impact of Health Problems on Independence

The impact of physical difficulties on adjustment and independence in adulthood was also examined. According to caregivers, only 20 subjects (29%) were receiving annual physical checkups from general practitioners or hospital consultants. A further 21 (30%) had received at least one physical examination during adulthood, while 29 subjects (41%) had never had any medical examination during their adult lives. Of the 20 subjects who received regular physical examination, one had required a heart transplant and 9 had hypertension which limited their physical activity and for which they received medication. The remaining 10 subjects received yearly cardiac monitoring, but their caregivers reported that their daily activities were not disrupted by cardiac problems or hypertension. Two subjects had received kidney transplants and were continuing to

attend hospital as outpatients, and a further 4 had kidney problems that required medication. One subject had received hospital treatment for a hiatus hernia during adulthood, and one suffered from rheumatoid arthritis which had required surgery. Bowel problems, such as prolapse of the rectum, hemorrhoids, diverticulitis, and constipation, were reported in 14 of the 20 subjects who received regular medical supervision, and 3 subjects had received minor gastrointestinal surgery. Significant dental problems were reported in half of this group.

Although not assessed systematically, additional health-related concerns expressed by the caregivers across the group as a whole included joint dislocation in the knees (9 subjects), complaints of aching limbs (30 subjects), and scoliosis (5 subjects). Painful periods or premenstrual tension were reported to be a problem for 36 of the 37 female subjects. Twenty subjects (29%) were regularly taking prescribed medication; of these, 6 received daily medication for raised blood pressure, 2 were taking medication following kidney transplants, and 1 adult was prescribed drugs for cardiac problems. Five subjects took medication for chronic constipation, and 1 took medication as a result of a hiatus hernia. One subject took antiepileptic drugs, 2 were receiving regular antidepressant medication, and 2 more received anxiolytic drugs. Thirty-six individuals (51%), 25 of whom were female, were considered overweight by their caregivers. No individual was reported to have had raised blood calcium levels at any time during adulthood.

Caregivers reported that hyperacusis caused significant problems for just under half of the group (46%), and supervision and reassurance were often required to help these subjects cope with sounds that caused distress. A further 52% of caregivers reported that, though certain sounds still caused some distress, subjects seemed better able to cope with exposure to these sounds than they had as children.

DISCUSSION

This study represents one of the largest investigations undertaken to date of the adjustment of adults with WS, in terms of self-help skills, independence levels, and occupational status. The present findings confirm many of the conclusions reached in earlier reports of functioning in adulthood [Plissart et al., 1994; Udwin, 1990]. People with WS continue to need substantial support and supervision in adulthood. Their limited fine-motor functioning, lack of persistence, and distractibility mean that most require prompting and supervision even when performing routine tasks such as washing and dressing. In addition, many parents were fearful about letting their son or daughter out without supervision. Visuoperceptual deficits, such as poor appreciation of speed and distance, could make crossing roads and negotiating busy streets particularly difficult, and noisy environments could cause distress due to hyperacusis. Parents were also very concerned that social disinhibition and overfriendliness could lead to exploitation. In the case of subjects who had left home, many parents also expressed concern about inadequate levels of supervision in residential homes and colleges. Occupational attainment was also very low, and very few of the sample were able to cope with employment. It was clear that individuals with WS often required considerable extra supervision in training and vocational settings.

Although the results reported here are generally in agreement with other studies of adults with WS, it is not clear whether the outcome for this group compares favorably or unfavorably with groups of people with other intellectually disabling genetic disorders. Among the few comparable studies, Dykens et al. [1992] reported Vineland Adaptive Behavior Scale scores for a group of 21 individuals with Prader-Willi syndrome (PWS) with a mean age $25\frac{7}{12}$ years. Despite having a similar degree of general cognitive impairment, the WS group scored relatively poorly on measures of communication and daily living skills, but rather better than the PWS group on measures of social independence. The scores attained by the WS group on the Daily Living Skills subdomains are comparable to those reported by Dykens et al. [1988] for a group of noninstitutionalized males with fragile-X syndrome. However, this group had a mean IQ of 50 and were also considerably younger (mean age, 14 years). Carr [1994] reported on the adaptive skills of a group of 41 adults with Down syndrome (DS) with a mean age of 21 years and a mean IQ below 50. Two thirds were rated as "independent" in their toileting, one to two thirds were independent in dressing, and about half in washing and bathing. These rates are strikingly similar to our own findings for individuals with WS, who were more cognitively able than the DS group.

In terms of living arrangements, Carr [1994] reported a similar proportion of her DS group living at home with their parents. These findings may reflect the limited range of alternative accommodations available for adults with learning difficulties. Greenswag [1987] investigated parents' ability to cope with adults with PWS who were living at home, and found that caregivers reported a large number of difficulties. In contrast, relatively few families of adults with WS who were still living at home were currently experiencing difficulties. This suggests that adults with WS may be rather easier to manage than adults with PWS. Nevertheless, our findings indicate that many families had reservations about their ability to cope with their son or daughter in the future, and caregivers recognized that accommodation with substantial levels of supervision was the most suitable residential arrangement for their son or daughter in the long term.

The breakdown of daytime occupations in this group is similar to the results presented by Putnam et al. [1988] and Carr [1994] for adults with DS, and by Waters et al. [1990] for adults with PWS. It is apparent that most adults with moderate and severe learning difficulties attend training and day centers. Employers and supervisors suggested that the difficulties in the workplace mostly arise from cognitive and personality characteristics known to be associated with WS, notably anxiety, social disinhibition, poor social functioning, visuomotor difficulties, and distractibility. Because of these characteristics, most individuals with

WS require considerable supervision in training and vocational settings, as well as continuing support and reassurance. Moreover, when making decisions about training and occupations for individuals with WS, their profile of strengths and deficits needs to be carefully considered. Routine manual tasks, such as stacking shelves, packing, or assembly-line work, which are typically considered suitable for people with learning difficulties, may not be suitable for many people with WS because of their visuomotor difficulties, lack of stamina, and tendency to tire easily.

Levels of independence were, in most cases, disappointingly low, and the results appear to confirm the previous conclusions of Bradley and Udwin [1989], Udwin [1990], and Morris et al. [1990], that in the case of individuals with WS, outcome in adulthood is limited more by behavioral and psychological factors than by physical difficulties. Only a minority of parents and caregivers reported that subjects were significantly impaired by chronic medical problems or had problems that required regular monitoring. Importantly, in those who received routine health checkups, serious medical problems were common. There are a number of documented cases of sudden death in this population, often as a result of cardiac disease, and all adults with WS should have their cardiac function, kidney function, and blood pressure checked at least once a year. It is clearly important that parents and health professionals do not underestimate the prevalence of health problems associated with WS, but it should also be noted that in the majority of cases in this study, caregivers did not consider that subjects had physical difficulties which significantly interfered with their functioning or which warranted medical investigation. Nevertheless, as the majority of subjects in this study were under 30, it is reasonable to suggest that progressive health problems may become more obvious in later life. Further information is required about the health risks associated with aging in this population.

In conclusion, the behavioral and psychological characteristics associated with WS (which include anxiety, distractibility, overfriendliness, lack of persistence, and poor gross and fine motor skills) appear to limit the level of self-care and community independence attained by affected individuals. Comparisons with other populations are difficult due to the different criteria used to define independence, and the use of different measures of cognitive functioning. Nevertheless, it appears that adults with WS need more support and supervision with self-care tasks and daily living skills when compared with other groups of adults with moderate or severe cognitive impairment. Expressive language abilities may disguise specific cognitive deficits, such as visuoperceptual and motor coordination difficulties, and often give the impression that individuals with WS are much more capable than they actually are. Awareness of the profile of strengths and weaknesses associated with the syndrome should enable supervisors, caregivers, employers, and families to provide appropriate residential and occupational opportunities for individuals to maximize their potential in a safe environment, which does not make excessive demands on their capabilities.

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